

# Case Presentation

From Pediatric M&M  
Fort Carson MEDDAC



# Case Presentation

12 month old male brought to clinic on 27 October for rectal fever 100-103° daily for 10 days.

Had been seen in ER the day before with temp 103°, labs were drawn.

Observed to be crying a lot, seems uncomfortable, decreased activity, no localized complaints.



# Case Presentation

- PMH – GI ‘bug’ about 2 weeks ago, sibs and parents also ill, recovered.
- PE - T=99.9°. NAD, non-toxic.
- +2 symmetrical, non-tender ant. Cervical lymph nodes.
- Otherwise unremarkable exam.



# Case Presentation

- LABS- UA normal
- Basic metabolic profile normal.
- WBC = 21.5, 63.6%N, 23.3% L.
- PLT= 545.
- H/H=12.4/36.1



# Case Presentation

- Questions up to now?



# Case Presentation

- No rash
- No redness or swelling of hands or feet.
- No erythema of lips, tongue, or eyes.



# Case Presentation

- Referred to Pediatric Cardiology to evaluate for possible Kawasaki Disease.



# Cardiology Evaluation

- Echocardiogram – “Some mild coronary artery ectasia bilaterally with normal coronary diameters and no visible aneurysms.”
- Admitted to Memorial Hospital
- Treated with IVIG





# Hospital Course

After treatment with IVIG 2 gm/kg fever rapidly defervesced

WBC on admission = 23.5

ESR = 58

PLT = 584

CRP = 5.46 (0.00-2.00)

Also received aspirin 160 mg QID

Discharged on 29 October with Pediatric Cardiology f/u in 1 week



# Follow-up

2 subsequent echocardiograms negative for aneurysms.

Seen again in ER 8 Nov for AGE from which he quickly recovered.

Has now moved from area and will continue Pediatric Cardiology f/u elsewhere.



## INCOMPLETE (ATYPICAL) KAWASAKI DISEASE

“INCOMPLETE” preferable to “ATYPICAL.”

These children lack sufficient clinical signs of disease to fulfill classic criteria, but do not demonstrate any really “atypical” features.

Incomplete type is more common in young infants than in older children.

Coronary artery aneurysms are also more common in young infants.

Lab findings of incomplete Kawasaki's are similar to those of classic disease.



# ANEURYSMS

Aneurysms rarely develop before day 10 of the illness.

Echocardiogram findings of perivascular brightness, ectasia, and lack of tapering of the coronary arteries in the acute stage of disease may represent arteritis before the formation of aneurysms.

Decreased left ventricular contractility, mild valvular regurgitation (typically mitral), and pericardial effusion may also be seen in the acute phase.



# Incomplete Disease

Incomplete Kawasaki Disease should be considered in:

All children with unexplained fever for  $\geq 5$  days associated with 2 or 3 of the principal clinical features

Any infant less than 6 months of age with fever  $\geq 7$  days' duration with any lab evidence of systemic inflammation (ESR, CRP, WBC, PLT) and no other explanation for the fever.

**These children should receive echocardiogram**



# Risk of Aneurysms

Duration of the fever – presumably reflecting the severity of the ongoing vasculitis – Is a powerful predictor of coronary artery aneurysms

Age  $\leq$  12 months

Male gender

Various scoring systems have been devised, but their imperfect performance necessitates the treatment with IVIG of all patients diagnosed with Kawasaki Disease.



# AHA Scientific Statement

*Diagnosis, Treatment, and Long-Term  
Management of Kawasaki Disease.*

CIRCULATION. 2004; 110:2747-2771



# Kawasaki Disease

## Definition:

An acute, self-limited vasculitis of unknown etiology that occurs primarily in infants and young children.

It is characterized by fever, bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy.

Coronary artery aneurysms or ectasia develop in 15 – 25% of untreated children and may lead to ischemic heart disease sudden death.





# Impact

In the U.S., Kawasaki Disease has surpassed Acute Rheumatic Fever as the leading cause of acquired heart disease in children


“Consultation with an expert should be sought anytime assistance is needed”

I.E. always have your patient seen by a Pediatric Cardiologist if you are seriously entertaining the diagnosis.



# Principal Clinical Findings

Fever persisting at least 5 days and the presence of at least 4 of the following:

1. Changes in the extremities: Acute – erythema of the hands and feet Convalescent – membranous desquamation of the fingertips
  2. Polymorphous exanthamata
  3. Bilateral painless bulbar conjunctival injection without exudate
  2. Changes in the lips and oral cavity – erythema and cracking of the lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosa
  3. Cervical lymphadenopathy ( $\geq 1.5$  cm diameter), usually unilateral
- 

## Nonpitting edema of hand



**Waseem, M. et al. Pediatrics in Review 2003;24:245-248**

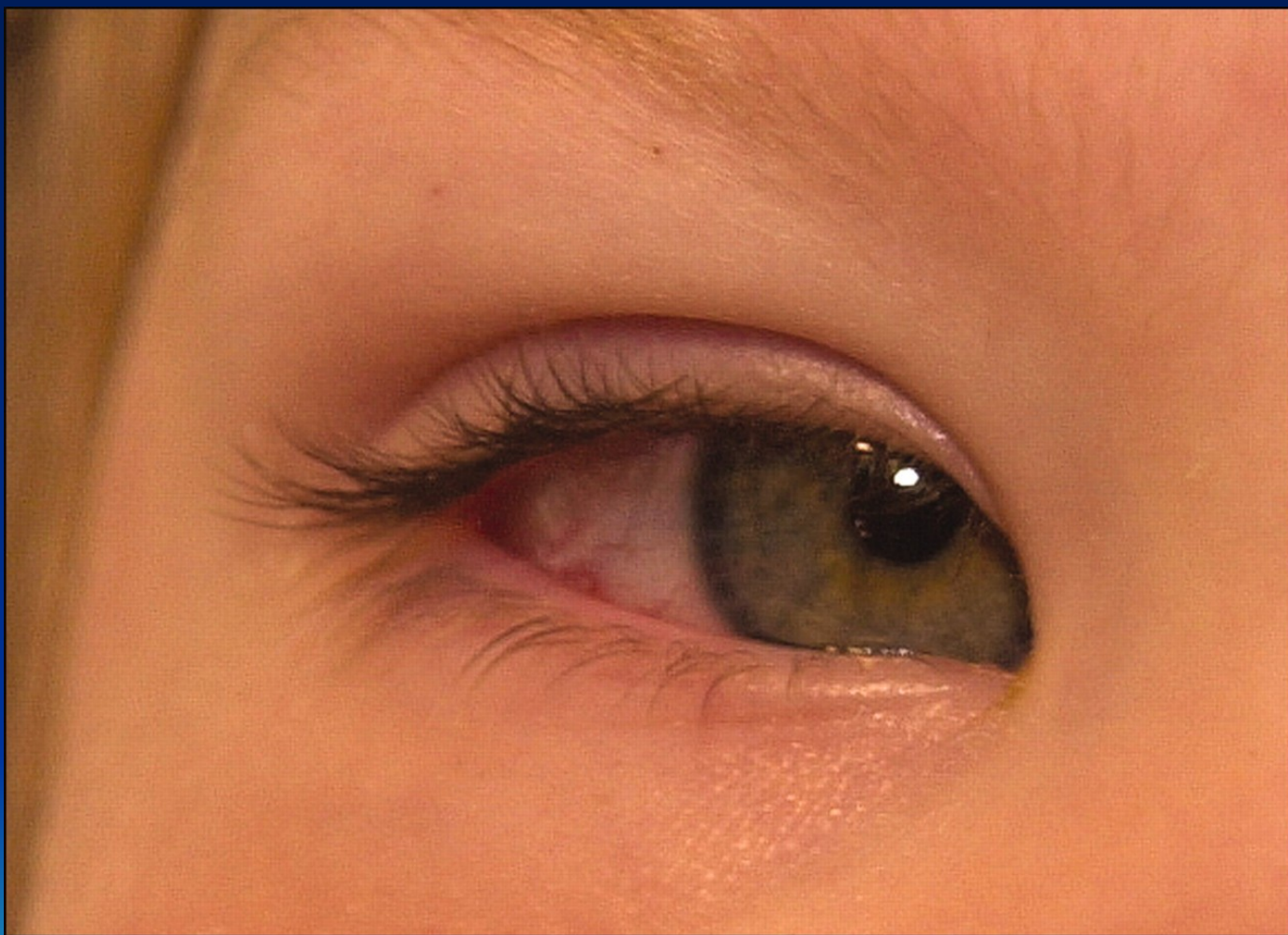
## Polymorphous red rash over extremities



**Waseem, M. et al. Pediatrics in Review 2003;24:245-248**



## Bulbar conjunctivitis without exudate



**Waseem, M. et al. Pediatrics in Review 2003;24:245-248**

## Red, fissured lips



**Waseem, M. et al. Pediatrics in Review 2003;24:245-248**

## Desquamating perineal rash



**Waseem, M. et al. Pediatrics in Review 2003;24:245-248**



# Characteristics Suggesting Disease Other Than Kawasaki

1. **Exudative** conjunctivitis
2. **Exudative** pharyngitis
3. Discrete intraoral lesions
4. Bullous or vessicular rash
5. Generalized adenopathy





# Supplemental Lab Data

1. Albumin  $\leq 3.0$  g/dl
2. Anemia for age
3. Increased AST, ALT
4. Platelets after 7 days  $\geq 450,000$
5. WBC  $\geq 15,000$
6. Urine  $\geq 10$  WBC/ hpf (sterile pyuria)

